

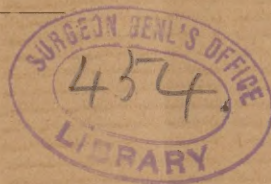
Hartzell (M. B.)

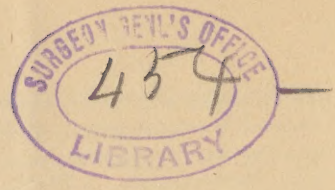
REPRINTED FROM
UNIVERSITY MEDICAL MAGAZINE,
May, 1890.

ACUTE CIRCUMSCRIBED CUTANEOUS ŒDEMA.

BY M. B. HARTZELL, M.D.

Instructor in Dermatology, University of Pennsylvania.





ACUTE CIRCUMSCRIBED CUTANEOUS ŒDEMA.

BY M. B. HARTZELL, M.D.

Instructor in Dermatology, University of Pennsylvania.

THE following case of what has been variously described as giant urticaria, acute circumscribed cutaneous, or angioneurotic œdema, has been under the writer's care during the past six months, and is reported because of the typical character of the lesions, the infrequency of the malady, and the scanty references to the subject in most text-books upon cutaneous diseases.

G. D., American; 40 years old; married; superintendent of a street-railway. First came under observation in the beginning of August, 1889, and presented at that time a large, ill-defined, rather soft, painless swelling, occupying the right cheek beneath the eye, and the entire right half of the upper lip. The skin over this swelling was normal in color, and except a slight sensation of stiffness in the affected parts, subjective symptoms were completely absent.

According to the patient's statement this lesion, which had existed about twelve hours, began some time during the previous night as a "small kernel" which rapidly enlarged, not uniformly in all directions but with great irregularity. This swelling rapidly reached its acme, and after remaining stationary for a short time as rapidly subsided, having completely vanished within twenty-four hours from its first appearance.

About five weeks later a new attack occurred; this time, however, the œdema was seated upon the left side of the face, covering the greater part of the cheek. A month later the left side of the face was again attacked; and during a period of five days swellings appeared upon various parts of the face, once completely closing the eyes for a short period. At this time the right and left halves of the tongue became greatly swollen in rapid succession.

Three weeks after this unusually severe outbreak swellings as large as an egg appeared upon the forehead and scalp. The mucous membrane of the larynx was attacked, and the patient's voice reduced to a whisper in consequence. This aphonia was, however, of brief duration, and was not accompanied by any disturbance of respiration.

A few days later an oblong, moderately well-defined swelling, measuring six inches in its long axis, three inches transversely, and elevated a half inch above the surrounding parts, appeared upon the flexor surface of the left forearm. This lesion, unlike those upon the face, was somewhat redder than the normal skin, and pitted slightly upon firm pressure. It may be remarked here that this was the only instance in which the œdema attacked parts of the cutaneous surface other than the face and scalp. During the last three months there have been but three œdematous lesions, and these have been quite insignificant in size compared with those occurring six months ago.

The disease has lasted three years or more, a month rarely passing without an attack. It is quite remarkable that these swellings always began some time during the night, the patient stating most positively that this has been the case without a single exception. They, likewise, always occurred singly; nor did a new one begin to appear before the old one had disappeared. It should also be noted that the patient frequently suffered from mild

attacks of urticaria, which sometimes occurred coincidently with the œdema of the face, but more frequently alone. The urticarial lesions presented nothing unusual in their appearance, were accompanied by only a moderate amount of itching and burning; and were never very numerous. Gastro-intestinal disturbances were completely absent both during the attacks of urticaria and cutaneous œdema. The most careful inquiry failed to reveal anything in the patient's diet or mode of life which could be regarded as having, even remotely, any etiological relationship with the cutaneous trouble; nor, so far as the patient could learn, had any member of his family, near or remote, ever suffered from a like disease.

In 1876, Milton¹ reported several cases of a malady characterized by the sudden occurrence of variously sized patches of œdema affecting the skin and mucous membranes, which ran an acute course, accompanied by slight or no subjective symptoms. For this malady he proposed the name giant urticaria. Later, Quincke,² Strübing³ and others reported cases of a similar kind under the name of acute circumscribed cutaneous or angioneurotic œdema.

In this connection a brief summary of the more prominent symptoms of this curious affection, as presented in the cases reported, may not be uninteresting. As a rule, the œdema affected chiefly the face, although in a considerable number of cases the trunk showed like lesions. The swellings are described usually as nut to egg sized; but in a few instances they reached much larger proportions, being the size of the fist, and even larger. In most cases no abnormal sensations were complained of beyond the tension occasioned by the swelling, although in a few itching, slight or severe, was present. Usually the skin preserved its normal color; occasionally it was pink or bright red. The duration of the individual lesions did not exceed two or three days, and was commonly much less, twelve to twenty-four hours being the length of time ordinarily required for the full development and involution of the œdema. The number of swellings existing at any one time was quite small, seldom more than two or three, and often they occurred singly, as in the writer's case. In several instances the mucous membrane of the tongue, soft palate, and larynx was attacked.

Milton and Strübing each report cases in which the cutaneous œdema was preceded by sudden and alarming attacks of dyspnœa, the true nature of which only became manifest after the occurrence of the swelling of the skin. In the case recorded by Strübing the dyspnœa reached such a degree that a fatal termination seemed imminent, and preparations were hastily made to perform tracheotomy, but the suffocative symptoms subsided as rapidly as they had come on.

In a large proportion of the cases nausea and vomiting, abdominal pain, or other evidences of gastro-intestinal derangement, accompanied the affection of the skin; and in some instances those who had suffered from the cutaneous œdema attended by vomiting and pains in the abdomen, subsequently had periodic attacks of gastro-intestinal disturbance without any cutaneous symptoms accompanying them. In a considerable number of the cases the attacks

¹ Edinburgh Med. Journal, 1876.

² Monatsheft f. praktische Dermatol., 1882.

³ Zeitschrift. f. Klin. Med., Bd. ix, p. 382.

recurred at regular intervals, and this was a fairly well-marked feature of the writer's case. Not infrequently urticaria of the ordinary type occurred, sometimes along with the œdema, but more often independently. It is to be remarked, however, that a fair proportion of the cases never suffered from any other cutaneous malady than the circumscribed œdema.

While acute cutaneous œdema presents many features which indicate a close relationship with urticaria, yet it possesses characters of its own which fairly entitle it to separate consideration. The sudden appearance and evanescent character of the individual lesions are strikingly like urticaria, but the great size of the swellings, the normal color of the skin, and the absence of itching and burning are points which distinguish it from that disease. Furthermore, the respiratory mucous membrane frequently shares in the morbid process, a complication very rare in urticaria. In this latter malady it is frequently possible to find the cause of the eruption in the ingestion of some particular article of food, such as shell fish, strawberries, etc. ; but in acute cutaneous œdema no such causal factor is to be discovered.

The etiology of the disease is involved in the utmost obscurity. But while without any knowledge of the immediate causes of the affection, the cases reported by Strübing and Osler make it evident that heredity plays an important rôle in its production. Strübing has reported an instance in which three members of the same family, the father and two children, were subject to attacks of acute œdema ; and more recently Osler has described an exceedingly interesting example of the hereditary character of this affection, in which one or more members of a family were affected in five generations.

The prognosis is extremely unfavorable, at least as regards a speedy cure, since in nearly all the recorded cases the attacks continued to recur at longer or shorter intervals over long periods of time, in one instance fifty years. When the mucous membranes become the seat of the œdema the affection is not devoid of danger, since a fatal termination from œdema glottidis has occurred.

As may be concluded from the foregoing, treatment exerts but little influence upon the disease. Milton reports favorable results from the use of tonics and laxatives. In my own case a saline laxative administered in small doses every morning, together with small quantities of salicylate of soda, seem to have diminished the frequency and severity of the attacks.

